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Case Report

Rhabdomyosarcoma of the biliary tract: a case report

Asmaa Kouadir*, Abderrahmane El Mazghi, Khalid Hassouni

Department of Radiotherapy, Hassan II University Hospital, Faculty of Medicine and Pharmacy of Fez, Sidi Mohamed Ben Abdellah University, Fez, Morocco

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***Correspondence:**
Dr. Asmaa Kouadir,
E-mail: kouadirasmaa@gmail.com

ABSTRACT

Rhabdomyosarcoma (RMS) of the biliary tract is a rare tumor that commonly arises from the common bile duct. The most common clinical symptoms are obstructive jaundice and abdominal pain. Although diagnosis is often difficult and is frequently made during surgery, diagnostic imaging techniques including ultrasound, computerized tomography scan, and magnetic resonance cholangiopancreatography remain useful in the diagnosis and evaluation of biliary tree anatomy. In order to improve prognosis, different rhabdomyosarcoma study groups have adopted multidisciplinary treatment approach. Herein we describe a case of three-year-old child with Embryonal rhabdomyosarcoma originating in the common bile duct who was treated with surgery, chemotherapy according to European soft tissue sarcoma group (EpSSG) protocol and adjuvant postoperative intensity modulated radiotherapy to surgical bed with 6 MV photons to a dose of 41, 4Gy in 23 fractions. One year and a half after the end of therapy, the patient is still disease free. Although Rhabdomyosarcoma of the biliary tract is a rare tumor, it should be considered in the differential diagnosis of patients who have obstructive jaundice and a cystic mass within the common bile duct. Once believed to be an incurable disease, the prognosis of patients with biliary rhabdomyosarcoma has improved with a multidisciplinary treatment approach.

Keywords: Biliary tract, Common bile duct, EpSSG protocol, Rhabdomyosarcoma, Treatment approach

INTRODUCTION

Rhabdomyosarcoma (RMS) of the biliary tract is rare in childhood but it is the most common cause of malignant obstructive jaundice in children. In contrary to other sites, it has been noted that the biliary tract contains tumors of only embryonal or botryoid histology.[1] Because of the scarcity of this disease, the majority of the pediatric literature consists of isolated case reports.[2,3] Here, we present a case of a male child with embryonal RMS originating in the common bile duct.

CASE REPORT

A 3-year-old male child with no known history of congenital abnormalities and no contributory family history, presented with complaints of abdominal pain associated to progressive obstructive jaundice evolving in a febrile context with asthenia and anorexia over one month period. On physical examination, the child was found to have jaundice, pallor, and a mild hepatomegaly; there was no adenopathy, no palpable masses, no splenomegaly, and no ascite.

Laboratory investigations showed an elevated conjugated bilirubin of 22mg/l with mildly elevated transaminases: ALT of 43 U/l and AST of 41 U/l, and negative hepatitis serologies.

Abdominal ultrasound (US) showed an aspect of a type III hepatic hydatid cyst, fistulizing into the common bile

duct (CBD) which measured 12mm of diameter, associated to a mild hepatomegaly.

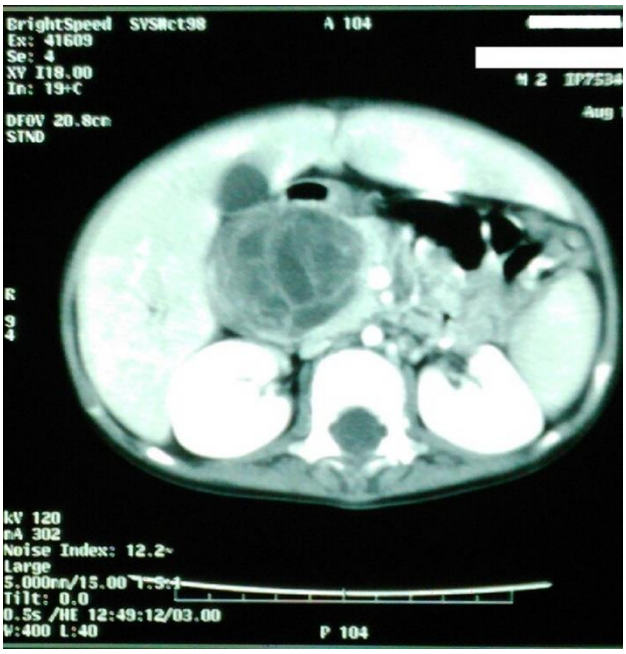

Figure 1: Post contrast CT scan at the level of the hepatic hilum showing dilated extrahepatic bile duct with Intraluminal heterogeneous mass.

Computerized tomography (CT) scan revealed a heterogeneous mass centered at the liver hilum with diffuse dilatation of the intrahepatic and extrahepatic bile ducts (Figure 1).

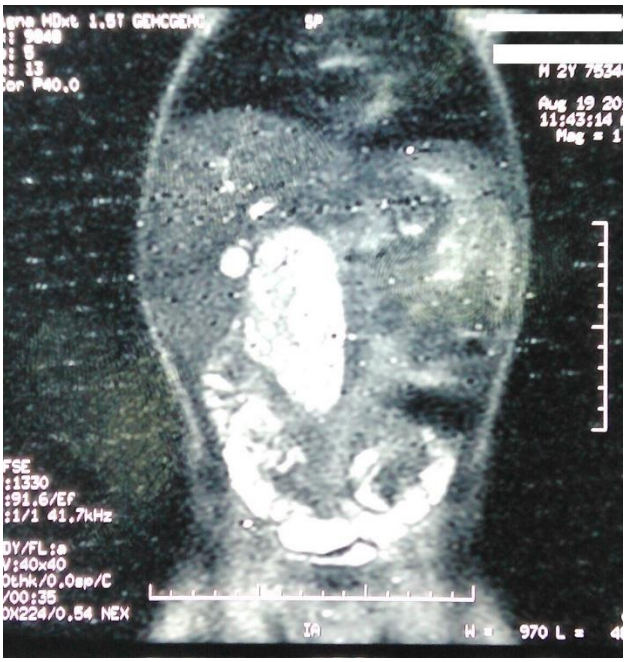

Figure 2: MRCP: Coronal T2-weighted image showing large heterogeneously enhancing mass responsible for voluminous common bile duct dilatation.

A subsequent magnetic resonance imaging (MRI) with 3D magnetic resonance cholangiopancreatography (MRCP) (Figure 2) confirmed the findings of a large 4.5 x 4.2 x 9 cm heterogeneously enhancing masse with mixed component (solid and liquid) situated in the hepatic hilum with inferior extension to the level of pancreatic head; this masse seemed to be within the lumen of the CBD and responsible for intrahepatic biliary dilatation. There were no lesions in liver parenchyma suggestive of metastasis; and no regional adenopathy was noted. Chest CT scan and Radionuclide Bone scan were also performed and showed no distant metastases.

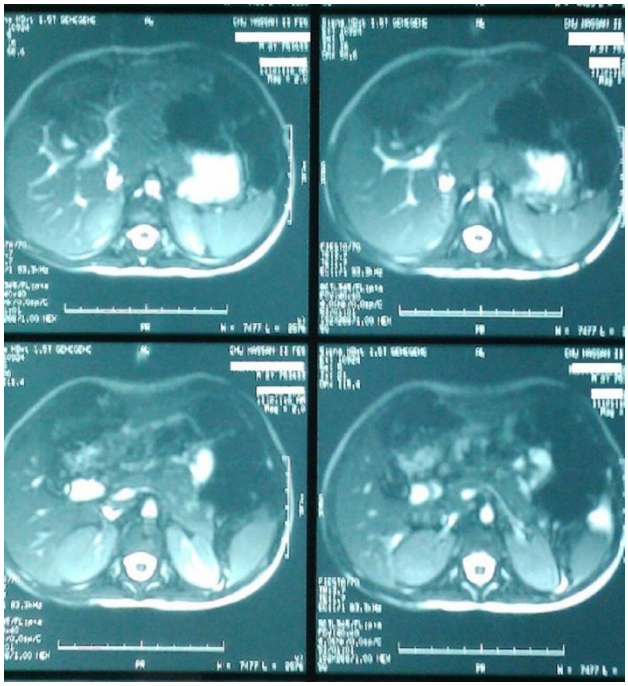

Figure 3: Abdominal MRI performed after three chemotherapy cycles: axial images showing no recurrence or residual tumor.

The patient subsequently underwent laparotomy with open exploration of CBD which revealed a 10cm x 4 cm myxomatous polypoidal tumor filling the lumen and adhering to the CBD wall. A cholecystectomy with complete resection of the common bile duct and a Roux-en-Y hepatico-jejunostomy was performed.

Histopathological examination revealed an embryonal botryoid RMS of the CBD affecting the entire choledochus till the common hepatic duct, and margins of the proximal resection were found invaded. There was no lymph node involvement

The case was classified into the IIa group of the intergroup rhabdomyosarcoma (IRS) clinical groups classification, and into subgroup E corresponding to a high-risk group according to the European soft tissue sarcoma group (EpSSG) risk stratification.

Twenty days after surgery, the child started adjuvant chemotherapy consisting of nine cycles associating Ifosfamide, Vincristine, and Actinomycin D (IVA), according to EpSSG RMS 2005 protocol.

After three chemotherapy cycles, abdominal RMI showed no recurrence or residual tumor (Figure 3). The patient was considered to have complete response. At week 17 after the start of chemotherapy, the child received an Intensity Modulated Radiotherapy (IMRT) which was delivered to surgical bed with 6 MV photons to a dose of 41, 4Gy in 23 fractions, while continuing chemotherapy cycles.

Imaging scans performed quarterly for one year and a half after the end of the therapy did not reveal any evidence of recurrence or metastatic disease.

DISCUSSION

Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma of childhood and occurs in a variety of anatomic locations.[1] Rhabdomyosarcoma of the biliary tract is rare, accounting for less than 1% of all pediatric RMS. It is a disease of young children (average age 3 years) with a slight male preponderance.[3,4] This tumor that mainly arises from the CBD may occur in any part of the biliary tree, resulting in symptoms of obstructive jaundice which is observed in 60%-80% of the cases. Clinical presentation may include other symptoms such as abdominal pain, emesis, fever, acholic stools, and decreased appetite.[1,4]

Laboratory finding of conjugated hyperbilirubinemia disproportionate to the mild elevation of hepatic transaminases is one of the diagnostic criteria. Diagnosis is often difficult and is frequently made during surgery.[1] Thus, before histopathological examination, most of reported cases in recent literature were misdiagnosed as choledochal cysts.[2,3]

Careful preoperative radiological examination remains nevertheless mandatory for the evaluation of biliary tree anatomy. While abdominal US often reveals dilated intra and extrahepatic bile ducts with a hyperechoic intraluminal mass within the ductal system, the appearance of biliary RMS on CT imaging can be highly variable. On MRI T1 and T2-weighted images, RMS commonly present as a T1 hypointense and T2 hyperintense mass.[5]

Macroscopically, the tumor occurs as an intraluminal biliary mass or cluster of grape-like masses. Microscopic characteristics of a polypoïd excrescence with a subepithelial density of cells are highly suggestive of embryonal RMS. Immunohistochemistry is very useful in the diagnosis. Actually, more than 95% of these tumors are positive for desmin. Moreover, because of its specificity for rhabdomyosarcoma, nuclear staining with myogenin is useful in the diagnosis.[6]

Table 1: Risk Stratification of European non-metastatic RMS Studies.

Post-surgical stage (IRS Group)	Sub groups	Pathology	Site	Size/Age	Node status	Risk group
I	A	Embryonal RMS	Any	≤ 5 cm and < 10 years	No	Low risk
	B	Embryonal RMS	Any	> 5 cm or ≥ 10 years	No	Standard risk
II, III	C	Embryonal RMS	Orbit, genito-urinary non-bladder or prostate tumor, non- parameningeal head and neck tumor	Any	No	Standard risk
	D	Embryonal RMS	parameningeal tumor, genito-urinary bladder or prostate, Extremities and "other site"	≤ 5 cm and < 10 years	No	Standard risk
	E	Embryonal RMS	parameningeal tumor, genito-urinary bladder or prostate, Extremities and "other site"	> 5 cm or ≥ 10 years	No	High risk
	F	Embryonal RMS	Any	Any	N1	High risk
I, II, III	G	Alveolar RMS	Any	Any	No	High risk
	H	Alveolar RMS	Any	Any	N1	Very high risk

Local treatment is one of the key aspects and the main prognostic factor in treatment of RMS. Both surgery and radiotherapy (RT) contribute to local control. Although previously authors have argued for aggressive surgery in the treatment of biliary RMS, this is not always feasible and can lead to significant morbidity. In a review of the

IRS I-IV data from 1972-1998, though most patients undergo initial surgical resection, gross total resection is accomplished in only 29%. Based on these findings, aggressive surgical resection in biliary RMS is discouraged. Therefore, most patients will receive postoperative radiotherapy.[1,3]

Table 2: Classification of tumor response to chemotherapy.

Tumor Response	Tumor volume reduction/ Initial tumor volume
Complete response	There is no measurable tumor
Good response	>2/3
Objective response	>1/3 and <2/3
Poor response	<1/3
Progressive disease	Volume increase of >1/3 or development of new lesions

Actually, RMS of the biliary tract is typically Embryonal or botryoid RMS which is more sensitive to therapy. Thus, radiotherapy (RT) is used in almost all biliary RMS patients (except Clinical Group I ERMS) to improve local control and outcome.[1,7] For EpSSG standard RT for RMS is delivered in once daily fractions to doses between 36 and 50.4Gy, varying according to the tumor histology, tumor response and IRS group.[7,8]

Although the benefit of RT is widely accepted, no consensus guidelines exist regarding specific radiation therapy parameters such as the use of IMRT in non-head and neck RMS. In a recent but limited retrospective review analyzing, inter alia, the impact of the use of IMRT on local control in pediatric patients with non-head and neck RMS, authors found no negative effect of the use of IMRT on local control. Yet, further analysis is required to evaluate the direct effects of this RT variable on treatment effectiveness as well as on treatment toxicity in non-head and neck RMS including biliary RMS.[8]

Regarding medical treatment, all patients with biliary RMS receive chemotherapy based on the risk grouping. In the children's oncology group COG, the risk stratification system incorporates pretreatment staging, the extent of disease after surgical resection (clinical group). For protocol purposes, patients are classified as low, intermediate, or high-risk. Moreover, the standard chemotherapy consists of vincristine, actinomycin-D and cyclophosphamide (VAC).

In the European soft tissue sarcoma group (EpSSG) the risk grouping, which depend on several prognostic factors, is divided into 8 subgroups which are further separated into low, standard, high and very high risk groups (Table 1). Rhabdomyosarcoma of the biliary tract is typically Embryonal RMS, thus, it could only be included in low, standard and high risk groups. On the other hand, ifosfamide, vincristine, and actinomycin-D (IVA) therapy is the standard treatment for RMS in European protocol. The associated lower gonadal toxicity justified the use of ifosfamide instead of cyclophosphamide.[7]

Furthermore, there is an important difference between COG and EpSSG treatment protocols which is the use of tumor response assessment using MRI or CT scan after three courses of chemotherapy. Tumor response to chemotherapy is classified according to the tumor volume reduction in relation to the initial tumor volume (Table 2). This response assessment aims to intensify chemotherapy in children with a tumor volume reduction below 50%. In COG studies, tumor response evaluation has not been utilized in order to guide therapy intensification and it has been often shown that response to therapy did not correlate with outcome.[7,9]

In our case, the patient received a total of nine chemotherapy courses with tumor response assessment using MRI after three courses according to EpSSG protocol. As the response to chemotherapy has been deemed complete, there was no chemotherapy intensification.

With the combination of therapies, survival has increased significantly in patients with biliary RMS and a recent study reported a survival rate higher than 75%, compared to 25% in 1970 in such patients.[10]

CONCLUSION

Although rhabdomyosarcoma of the biliary tract is a rare tumor, and its preoperative diagnosis is extremely difficult. It should be considered in the differential diagnosis of patients who have obstructive jaundice and a cystic mass within the CBD. With advances in the combined treatment of surgery, radiotherapy and chemotherapy, the prognosis of biliary RMS has become much better and the chances of long term survival have improved.

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